

# Radiological Manifestations of Kimura’s Disease: A Case Report

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## ABSTRACT

Kimura disease is a rare and non malignant soft tissue disorder characterised by asymptomatic swelling, predominantly in the craniofacial region. It is commonly associated with elevated eosinophil counts and systemic manifestations. This case study covers a 45-year-old male who presented with substantial right parotid oedema that had developed over five years. He was referred to the Radiology department for Magnetic Resonance Imaging (MRI), which revealed an enlarged superficial lobe of the right parotid gland, hypointense on T1 and hyperintense on T2/FLAIR. Multiple enlarged lymph nodes with diffusion restriction were observed at the right levels Ib, II, and III, with the largest at level II on the right-side. The bilateral lacrimal glands were prominent, with postcontrast enhancement. The postcontrast T1-weighted MRI showed tubular signal-intensity voids, likely representing fast-flowing vascular structures. When an individual of Asian descent presents with a subcutaneous mass that is either partially or poorly delineated, exhibiting high signal intensity on T1- and T2-weighted imaging, accompanied by uniform enhancement, adjacent subcutaneous oedema, and internal flow voids, it is imperative to consider Kimura’s disease, particularly in the presence of peripheral eosinophilia. This article focuses on the essential aspects of the disease and how the diagnosis is determined based on MRI results, supported by histopathologic features.

**Keywords:** Asian descent, Flow voids, Parotid oedema, Vascular channels

## CASE REPORT

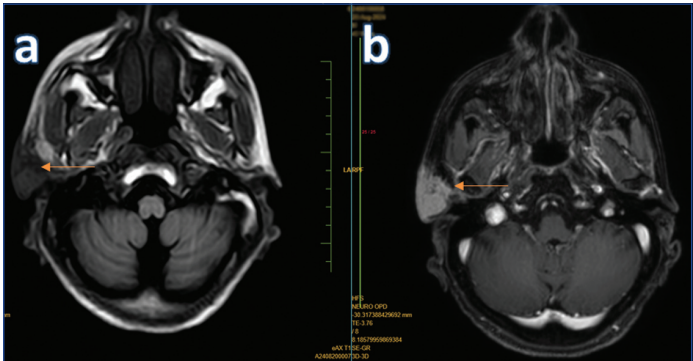
A 45-year-old male presented to the surgical department with the major complaint of right parotid swelling for over five years, which had gradually increased in size. The patient experienced an episode of loss of consciousness three days ago, prompting him to visit the hospital. He had a history of intermittent hypoglycemic episodes. The patient was hypotensive and denied any history of fever or weight loss. The underlying skin was affected over the right parotid area, as shown in [Table/Fig-1]. There appeared to be a skin patch near the jawline and cheek with lighter pigmentation than the surrounding skin. The texture in the affected area seemed dry and scaly compared to the rest of the face. Upon thorough examination, the pulse rate was determined to be 78 beats per minute, the blood pressure recorded was 80/40 mm Hg, and the respiration rate was ascertained to be 22 breaths per minute. The patient presented with no fever.



**[Table/Fig-1]:** Clinical image of the patient showing the skin changes in the affected region of right parotid.

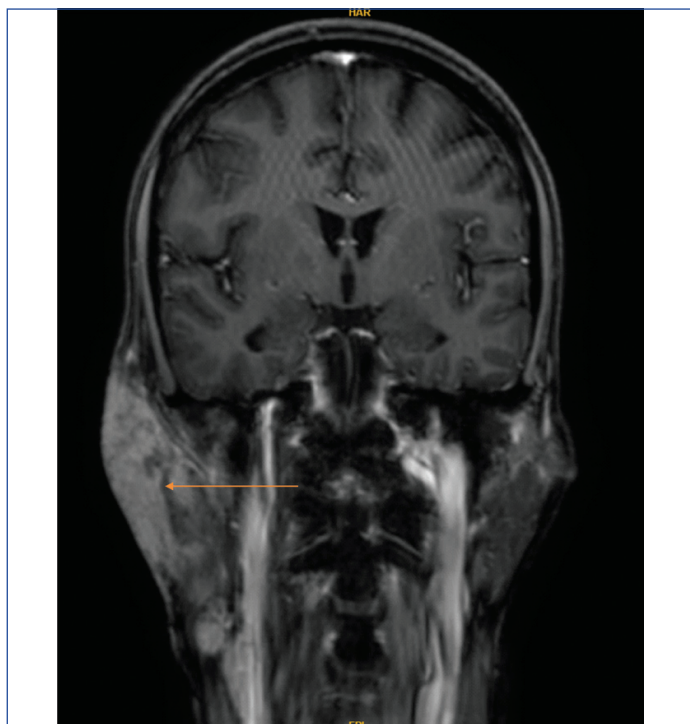
Laboratory studies indicated a haemoglobin level of 10 gm/dL (normal value-13.8 to 17.2 g/dL), Packed Cell Volume (PCV) of 31.5% (normal value-40.7 to 50.3%), an Mean Corpuscular Volume (MCV) of 73 fL (normal value-80 to 100 femtoliters (fL)), Red Blood Cell Distribution Width (RDW) of 15.2% (normal value-11.5 to 14.5%), an increased White Blood Cell (WBC) count of 16,700/cumm (normal value-4,000 to 11,000 cells per microliter (cells/ $\mu$ L)), and an absolute eosinophil count of 617/cumm {normal value-0 to  $0.5 \times 10^9$ /L (eosinophils per liter of blood)}. Serum lipase levels were elevated at 96.9 IU/L (normal value-10 to 140 units per liter (U/L)). Because the patient suffered from recurring hypoglycemic crises, an HbA1C test was performed, which revealed a level of 4.9% (normal value-Less than 5.7%).

The patient was advised to undergo an MRI of the brain as part of the work-up, which indicated that the superficial lobe of the right parotid gland was enlarged, appearing hypointense on T1, as shown in [Table/Fig-2a], hyperintense on T2/FLAIR, with diffusion on Diffusion-Weighted Imaging (DWI) and homogeneous postcontrast enhancement measuring 31×20×66 mm (AP×TR×CC), as shown in [Table/Fig-2b,3]. Multiple enlarged lymph nodes with diffusion restriction were observed at the right Ib, II, and III levels, with the largest measuring 17×13 mm at level II on the right-side, as shown

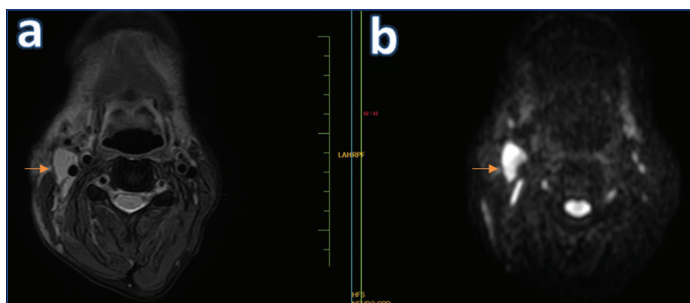


**[Table/Fig-2]:** Axial MRI brain T1 (a) and T1 weighted post contrast; (b) Images showing parotid gland appearing hypointense on T1 and homogeneously enhancing in post contrast image.

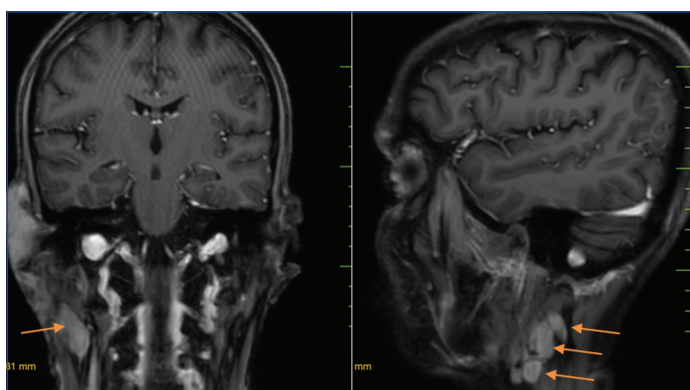
in [Table/Fig-4,5]. The bilateral lacrimal glands were prominent, with postcontrast enhancement. The postcontrast T1-weighted MR image showed tubular signal-intensity voids, probably representing fast-flowing vascular structures, as shown in [Table/Fig-6]. The patient was advised to undergo fine needle aspiration cytology. Histopathological analysis revealed abundant vascular proliferation (hyperplasia), as shown in [Table/Fig-7].



**[Table/Fig-3]:** Coronal post-contrast MRI brain image showing homogenous enhancement of right parotid gland.

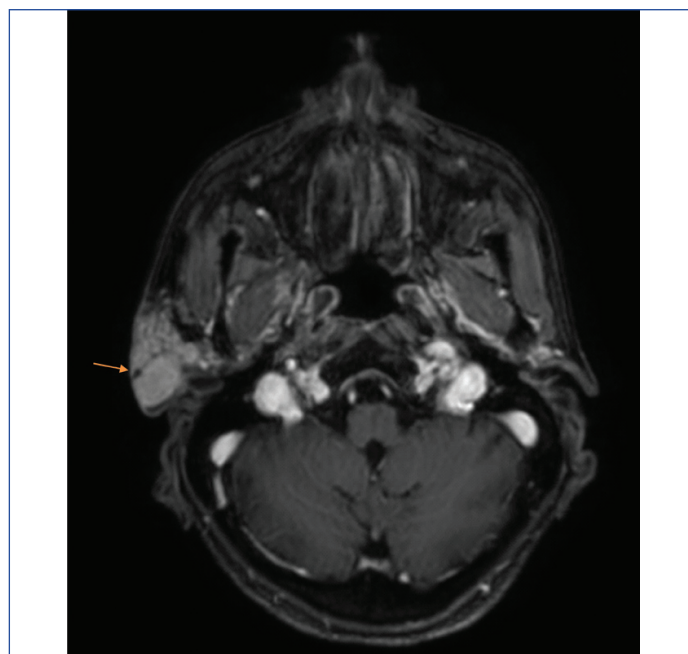


**[Table/Fig-4]:** Axial MRI brain T2: (a) and DWI stir; (b) showing largest enlarged lymph node in level II.

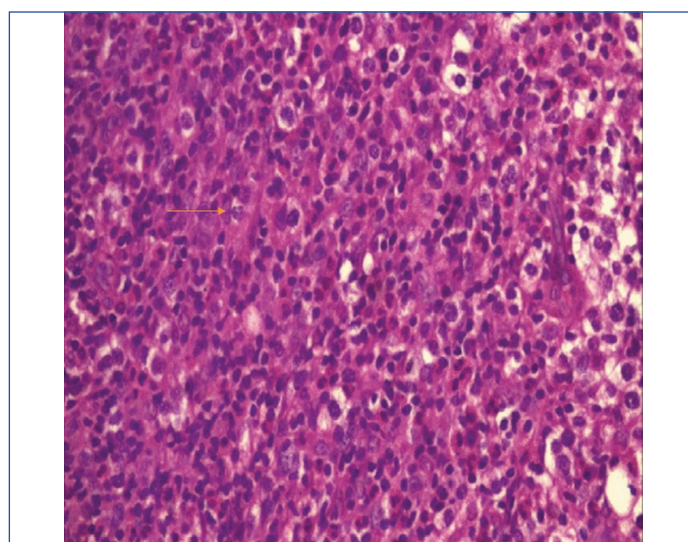


**[Table/Fig-5]:** T1 post-contrast coronal and sagittal sections of the brain showing multiple enlarged lymph nodes, largest on the right-side.

In this case, Kimura disease was established based on the patient's presentation of several enlarged lymph nodes, diagnostic MRI findings, and histological examination. The patient was instructed to attend the Outpatient Department (OPD) to plan the treatment after the diagnostic imaging, but he did not arrive, and thus no treatment was scheduled.



**[Table/Fig-6]:** Axial MRI brain T1 post-contrast image showing tubular signal-intensity voids, representing fast-flowing vascular structures.



**[Table/Fig-7]:** Histological slide showing vascular hyperplasia (H&E stain, 40x).

## DISCUSSION

Kimura's illness is a rare chronic inflammatory disease that mostly affects young Asian males [1], with males being affected three times more than females [2]. It usually appears as painless cervical adenopathy or subcutaneous tumours in the head and neck, primarily affecting the salivary glands and lymph nodes [3]. The axilla, popliteal area, groin and forearm are less common sites of involvement [4,5]. The most typical clinical findings are asymptomatic enlargement of the salivary glands and lymphadenopathy. The presence of peripheral blood eosinophilia and elevated serum IgE concentrations suggests that the illness is immunologically caused. Atopic characteristics include IgE deposits in germinal centers, peripheral eosinophilia, high blood IgE levels and the development of mast cells [6].

The anatomical regions frequently implicated in this condition include the periauricular area, the epicranium, the orbit and the eyelids, which are all significant sites of involvement that warrant attention in clinical assessments [7]. In individuals who are not of Asian descent, it is noteworthy that the salivary glands are rarely affected by this condition; however, there are instances in which the lacrimal glands may also be involved, thus highlighting the variability in clinical presentation among different ethnic groups [8]. The clinical progression of Kimura disease is characterised by a tendency toward



advancement over time, and a prominent challenge associated with the management of this disease is the recurrent nature of its manifestation, which poses complications for effective treatment strategies. The infrequency of this ailment renders the establishment of a conclusive treatment protocol a challenging endeavor. This condition is considered benign and exhibits a particular inclination toward younger patients. Consequently, the application of irradiation is infrequent due to apprehensions regarding the potential development of secondary malignancies [9].

Peripheral blood eosinophilia (10%-70%) and increased serum IgE levels (800-35,000 IU/mL) are common [10] and may vary throughout the disease's progression. Numerous studies have identified a significant correlation between the dimensions of lesions and the presence of eosinophilia; specifically, it has been observed that an increase in lesion size corresponds with elevated levels of eosinophilia in the peripheral bloodstream [10,11]. The serum levels of eosinophil cationic protein may serve as an additional measure of disease activity.

Computed Tomography (CT) scans demonstrate clearly defined nodular or plaque-like infiltrative lesions located within the subcutaneous tissue, which are concomitant with lymphadenopathy. The preponderance of these findings is observed in proximity to the major salivary glands, with a particular emphasis on the parotid gland [12]. MRI exhibits varying signal strength, with enhancement patterns ranging from mild to intense and homogeneous to heterogeneous. MR scans may reveal serpentine regions of signal intensity voids, indicating the presence of vascular structures within the lesions [13].

Choi JA et al., found dot-like or tubular signal intensity voids inside the mass in all nine instances of Kimura disease affecting the upper extremities in an MRI examination, which were also noted in present case [14]. Kimura disease lesions appear diverse, hypointense, and sometimes slightly hyperintense on T1 and T2 weighted imaging [15]. Takahashi S et al., indicate that these lesions exhibit heterogeneous enhancement and are influenced by significant fibrotic tissue [5]. The study demonstrates that contrast enhancement in the parotid gland is caused by differences in fibrosis and vascular proliferation, as well as by the presence of a soft-tissue tumour, lymphadenopathy, eosinophil and lymphocyte infiltration, and dense fibrotic tissue.

A 36-year-old male presented with recurring, painless swelling in the right postauricular area. The MRI revealed a well-defined, lobulated mass in the periparotid region, with isointensity on T1-weighted images and heterogeneous hyperintensity on T2-weighted imaging. Postcontrast sequences showed strong, homogeneous enhancement, indicating the lesion's hypervascularity-a hallmark of Kimura disease. Additionally, DWI revealed limited diffusion within the lesion, validating the diagnosis [16]. Another study of 28 individuals with Kimura disease found that MRI and CT imaging often reveal ill-defined lesions in the head and neck region, with the parotid and periparotid regions being the most commonly affected. These lesions frequently show homogeneous enhancement upon contrast injection, indicating their hypervascular character [17].

The differential diagnosis for KD in the head and neck includes parotid tumours, lymphoma, metastatic tumours, malignant tumours of cutaneous or subcutaneous origin, Tuberculosis (TB), and angiolymphoid hyperplasia with eosinophilia [5,12]. It is vital to understand the distinctions between Kimura disease and Angiolymphoid Hyperplasia with Eosinophilia (ALHE), as shown in [Table/Fig-8].

Treatment for Kimura disease includes surgical excision and regional or systemic therapy [18]. Surgical excision is the preferred first-line therapy for isolated lesions, as it allows for the complete removal of the affected area. When surgical intervention is not possible or additional therapy is necessary, radiation therapy can be employed

Kimura disease	Angiolymphoid hyperplasia with eosinophilia
Common in 2 <sup>nd</sup> -6 <sup>th</sup> decade	Common in 3 <sup>rd</sup> -4 <sup>th</sup>
Males>Females	Females>Males
Salivary gland involvement (40%) and Lymph node involvement (70%) is reported	Involvement of bone, muscle, blood vessels and nerve
Peripheral eosinophilia is seen in 90% of cases	Peripheral eosinophilia is seen in 20% of cases
Serum IgE is elevated	Usually normal
[Table/Fig-8]: Differences between Kimura disease and angiolymphoid hyperplasia with eosinophilia.	

as a viable option. To treat more widespread or resistant cases, systemic immunosuppressive medication, such as corticosteroids or cyclosporine, may be utilised. Emerging medications, such as dupilumab, a monoclonal antibody that targets interleukin pathways, have shown promise, with some patients experiencing excellent outcomes [19]. These options emphasise the significance of personalised treatment strategies tailored to the severity and nature of the lesion.

CONCLUSION(S)

This case report highlights the radiological study of Kimura disease in a 45-year-old man. The illness was identified and differentiated from other comparable disorders using an MRI. The results showed enlarged lymph nodes and soft-tissue tumours with defined borders. Early detection and effective treatment, such as corticosteroid medication or surgical excision, are critical for alleviating discomfort and preventing recurrence. This case underscores the importance of considering Kimura disease in the differential diagnosis of patients with painless subcutaneous lumps and lymphadenopathy, especially in endemic areas. Further study is encouraged to better understand the clinical and radiological manifestations.

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#### PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Oct 10, 2024
- Manual Googling: Jan 09, 2025
- iThenticate Software: Jan 11, 2025 (12%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

#### AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: Oct 09, 2024

Date of Peer Review: Dec 17, 2024

Date of Acceptance: Jan 14, 2025

Date of Publishing: Mar 01, 2025